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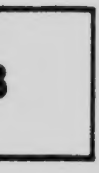

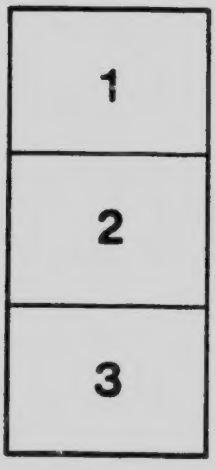

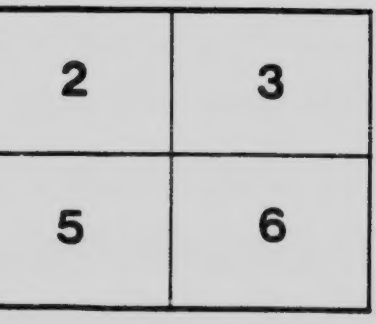

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On the Differentiation of Two Forms of Congenital Dextrocardia

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ON THE DIFFERENTIATION OF TWO FORMS OF CONGENITAL DEXTROCARDIA.

By M. E. Abbott and J. C. Meakins,
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The heart may lie to the right of the median line in the thorax from a variety of causes, congenital and acquired. Three distinct types may be recognized. In the first of these, to which the name of *dextro-versio cordis*, may be applied, the alteration in position is due to extraneous causes, the organ being pushed, pulled or dislocated to the right from pathological conditions outside of itself, such as effusions or new growths in the left pleura or lung. In these cases the causative factor causing the displacement, is usually acquired or post-natal disease, but in some rare instances it may be of congenital origin, as for instance, in great enlargement of the left lung from congenital cystic disease. In these cases although the heart is displaced to the right the apex still points to the left and is usually felt during life not further to the right than the right sternal border.

In the other two types the condition consists, not in a simple pushing over of the heart to the right, but in its congenital malposition from causes acting within the heart itself. These are the cases of true congenital dextrocardia. In these the heart is situated to the right of the median line, and the apex points to the right, the apex beat during life being situated some distance to the right of the right sternal border. We owe the clear differentiation of these two forms of congenital dextrocardia to the studies by Kuchenmeister,[1] in a casuistic contribution on 125 cases, and more recently by Nägel[2]. The two forms of true congenital dextrocardia may be distinguished as types A and B.

In *type A* there exists a true transposition of the heart. Here all the cavities with their afferent and efferent vessels are reversed in relation to each other, so that a mirror-picture of the normal heart results. This condition is practically always associated with a more or less complete transposition of the other

[1] Kuchenmeister, Leipzig, 1883.
[2] Nägel, Deut. Arch. f. klin. Med. vol. XCVI, p. 552.

viscera, a *situs inversus* existing. In these cases the apex which points to the right is formed by the ventricle that normally lies on the left side, namely, that receiving blood from the left (pulmonic auricle). So also the chambers here occupying the left side of the heart are those lying normally on the right and receiving blood from the venae cavae. This condition cannot be regarded as pathological and is the result, not of an arrest of development, but probably of the inversion of the embryo in its relation to the primitive yoke-sack.

Type B. In this, the other form of congenital dextrocardia, the heart is not reversed upon itself, but is simply placed more or less in the right side of the thorax, and its apex points to the right and is formed by the right, that is the caval ventricle. The auricles maintain their normal relation, that on the right side receiving the venae cavae, and that on the left side receiving the blood from the pulmonary veins, which passes into the ventricle lying on the left side of the heart, and giving off the aorta. These cases are plainly due to arrest of development in the early embryonic stage, in which the heart lies chiefly on the right side, and the apex is formed of the right side of the common ventricle, which at that time preponderates over the left. They are practically always associated with marked cardiac defects, such as absence or defect of the septa, transposition of the arterial trunks, pulmonary hypoplasia, etc., a fact which gives additional proof of their origin in an arrest of development. These conditions are pathological and place the subjects of this type of dextrocardia in the class of those suffering from congenital cardiac disease.

The differentiation of these two types is excellently brought out in a set of diagrams by Nägel in his article on this subject, which are reproduced in Figures 1 to 5. We have added in

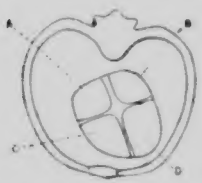

FIG. 1.—NORMAL HEART. A, CAVAL AURICLE; B, PULMONARY VEINS AURICLE; C, RIGHT VENTRICLE (CAVAL BLOOD VENTRICLE); D, LEFT VENTRICLE (PULMONARY BLOOD VENTRICLE).

Fig. 1. *Diagram of Normal Heart.* The apex points to the left, and is formed by the left (pulmonic blood) ventricle.

FIG. 2.—EMBRYONIC HEART. A, CAVAL AURICLE; B, PULMONARY VEINS AURICLE; C, RIGHT VENTRICLE (CAVAL BLOOD VENTRICLE); D, LEFT VENTRICLE (PULMONARY BLOOD VENTRICLE).

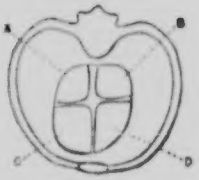

Fig. 2. *Diagram of Embryonic Heart.* The apex points to the right, and is formed by the right (caval blood) ventricle.

FIG. 3.—MIRROR-PICTURE OF NORMAL HEART. A, CAVAL AURICLE; B, PULMONARY VEINS AURICLE; C, RIGHT VENTRICLE); D, LEFT VENTRICLE (PULMONARY BLOOD VENTRICLE).

Fig. 3. *Diagram of Mirror Picture of Normal Heart.* The apex points to the right and is formed by the left (pulmonic) ventricle, and the caval and pulmonic auricles lie in reversed relations on the left and right sides respectively.

FIG. 4.—PURE CONGENITAL DEXTROCARDIA DUE TO EMBRYONIC ARREST. A, CAVAL AURICLE; B, PULMONARY VEINS AURICLE; C, CAVAL BLOOD VENTRICLE; D, PULMONARY BLOOD VENTRICLE. NOTE SIMILARITY TO NO. 2.

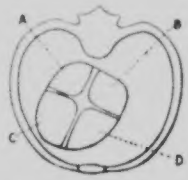

Fig. 4. *Diagram of Heart in Pure Congenital Dextrocardia due to Embryonic Arrest* of development. Note that the apex points to the right, and is formed by the right (caval blood) ventricle, as in Fig. 2.

FIG. 5.—CONGENITAL DEXTROCARDIA IN SITUS INVERSUS (MIRROR-PICTURE). A, CAVAL AURICLE; B, PULMONARY BLOOD AURICLE; C, CAVAL BLOOD VENTRICLE; D, PULMONARY BLOOD VENTRICLE.

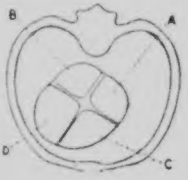

Fig. 5. From a case of *Pure Congenital Dextrocardia in Situs Inversus (mirror-picture dextrocardia).* Note that the apex points to the right and is formed by the left (pulmonic blood) ventricle, as in Fig. 3.

From Nägel's Pure Congenital Dextrocardia, *Deut. Arch. f. klin. Med.* XCVI. P. 572.

Fig. 6.—Grunmach's Case of Pure Congenital Dextrocardia (Arrest in Embryonic Stage). E, Vena Cava Superior; F, Ventriculum; G, Aorta; H, Ventriculum Sinistrum.

Fig. 6. Picture of *Grunmach's case of Congenital Dextrocardia* without situs Inversus *due to arrest of development*. Note that the apex points to the right but is formed by the right (caval blood) ventricle as in Figs. 2 and 4. See his article Berlin klin. Woch. 1890, p. 22.

The auricles are designated "caval" and "pulmonary" (according to the veins they receive), instead of right and left, in order to show the difference in situation in the mirror-picture cases, from that of the usual type of pure congenital dextrocardia due to embryonic arrest(Grunmach's case).

Figure 6, the sketch of a case figured by Grunmach of the type of dextrocardia due to arrest of development in which the apex points to the right and is formed of the right (caval) ventricle, and in which grave defects of development are associated.

It is seen from the above statement that these two types, A and B, of congenital dextrocardia are of very different clinical import, that of Type A (mirror-picture Dextrocardia) *in situs inversus*, occurring in otherwise normal and perfectly healthy individuals, and being of interest purely from the standpoint of a natural anomaly. That of type B, due to embryonic arrest, on the other hand, belonging to the gravest and most complicated forms of congenital cardiac disease, and almost always certainly associated with the symptom-complex of cyanosis and clubbing.

The fact that the electro-cardiogram supplies a definite diagnostic method for distinguishing the dextrocardia of *situs inversus* from the normal condition and also from the dextrocardia due to embryonic arrest of development is of great interest.

The electro-cardiograph differentiates the cases of mirror-picture dextrocardia (Type A) from the normal condition and from the dextrocardia due to embryonic arrest (Type B). In the normal heart the Sino-auricular node is situated at the junction of the Superior Vena Cava (*dextra*) and the right auricle, while in mirror-picture dextrocardia, the Sino-auricular node is placed at the junction of the Superior Vena Cava (*sinistra*), thus there is in the latter a complete inversion of the sino-auricular node in the relation to the body in general. In the normal electro-cardiogram, Lead I is taken from the right arm to the left arm.

similar lead is taken in a case of mirror-picture dextrocardia, electro-cardiographic tracing in the latter condition will be exact inverse of the normal case. This can be further proven reversing the current in the normal case from the left arm to right arm, when an inverted tracing will be obtained, whilst a case of mirror-picture dextrocardia the lead from the left to the right arm will give a tracing similar to the right arm left arm lead in a normal case. Leads II and III are reversed not inverted.

In a case of dextrocardia of congenital defect, in which the lies to the right, and is formed by the right ventricle, (Type a definite electrocardiographic tracing is obtained. In such case there is a marked preponderence of the S summit in Lead with no or very little R summit, whilst in Lead III the R summit is very much exaggerated while the S summit may be absent.